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Leukaemia Section

Mini Review

t(11;14)(q13;q32)

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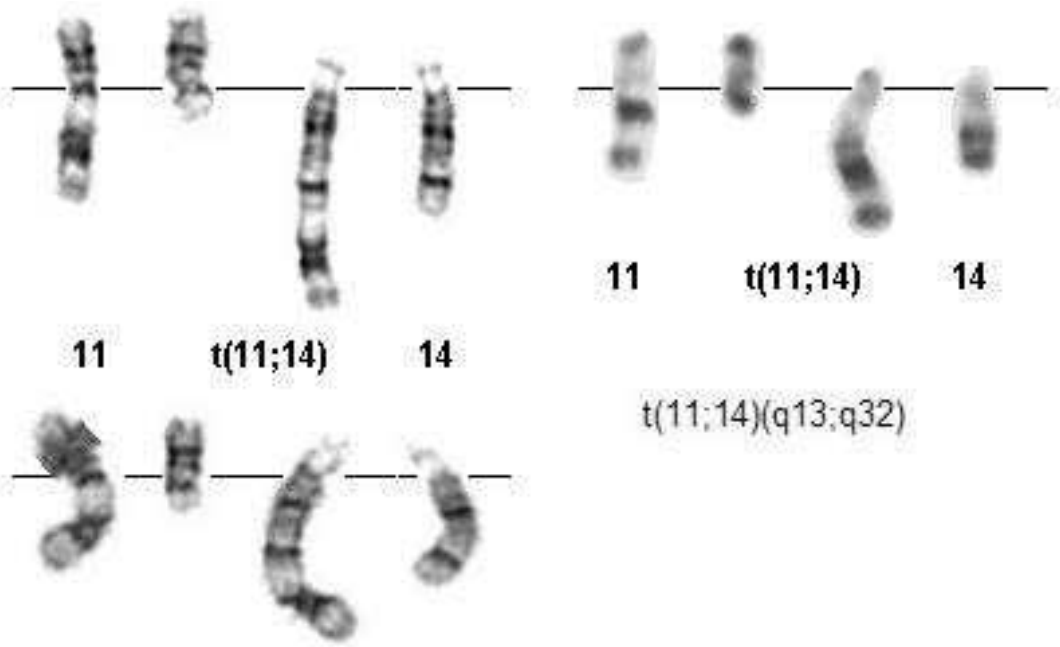

t(11;14)(q13;q32) G-banding (left) - Courtesy Diane H. Norback, Eric B. Johnson, Sara Morrison-Delap Cytogenetics at the Waisman Center and R-banding (right) - Editor.

Note: t(11;14) is mainly found in mantle cell lymphoma, but also in B-prolymphocytic leukaemia, in plasma cell leukaemia, in splenic lymphoma with villous lymphocytes, in chronic lymphocytic leukaemia, and in multiple myeloma, herein briefly described; all these diseases involve a B-lineage lymphocyte

Clinics and pathology

Disease

Mantle cell lymphoma.

Phenotype / cell stem origin

B-cell non Hodgkin lymphoma of the low to intermediate grade.

Epidemiology

Annual incidence $5/10^6$; median age: 65 yrs.

Clinics

Advanced disease.

Prognosis

Median survival: 3 to 4 yrs.

Disease

B-prolymphocytic leukaemia.

Phenotype / cell stem origin

Chronic lymphoproliferative disorder affecting mature B-cells.

Epidemiology

Rare disease; median age 70 yrs.

Clinics

Patients often present with advanced stage disease.

Prognosis

Median survival: 3 yrs.

Disease

Plasma cell leukaemia.

Phenotype / cell stem origin

Proliferation involving plasma cells.

Epidemiology

Rare disorder.

Prognosis

Median survival is less than a yr.

Disease

Splenic lymphoma with villous lymphocytes.

Phenotype / cell stem origin

Chronic B-cell lymphoproliferation.

Epidemiology

Rare disorder; median age: 70 yrs.

Clinics

Relatively benign clinical course.

Prognosis

80% 5-yr survival.

Disease

Chronic lymphocytic leukaemia.

Phenotype / cell stem origin

Chronic B-cell lymphoproliferation.

Epidemiology

Annual incidence $30/10^6$; median age: 60-80 yrs.

Clinics

Often a slow evolutive disease.

Prognosis

Highly variable according to the staging: from staging A: survival not reduced compared to age matched population, to staging C: median survival of 2 yrs.

Disease

Multiple myeloma.

Phenotype / cell stem origin

Malignant plasma cell proliferation (terminally differentiated B-cell).

Epidemiology

Annual incidence: $30/10^6$; median age: 60 yrs.

Prognosis

Median survival: 3 yrs.

Cytogenetics

Cytogenetics, morphological

t(11;14) has earlier been thought to be the hallmark of the mantle cell lymphoma; actually, the frequency of t(11;14) is: 50-70% in mantle cell lymphoma, 10-20% in B-prolymphocytic leukaemia, in plasma cell leukaemia, and in splenic lymphoma with villous lymphocytes, and 2-5% in chronic lymphocytic leukaemia, and in multiple myeloma.

Cytogenetics, molecular

In particular interphase cytogenetics, are relevant in these diseases with an usually low mitotic index.

Additional anomalies

Sole anomaly in only 10% of cases; part of a complex karyotype in 2/3 of cases; numerous recurrent anomalies found conjointly (which is the primary?), particularly: +3, +7, del(9p), +18, +mar, found in about 10% of cases each; other: del(1p), del(6q), del(7q), -8, +12, del(13q), del(17p).

Variants

Three way complex t(11;14;Var) exist and showed that the crucial event lies on der(14).

Genes involved and Proteins

BCL1

Location: 11q13

DNA / RNA

5 exons.

Protein

Encodes the cyclin D1; role in the cell cycle control: G1 progression and G1/S transition.

IgH

Location: 14q32

Results of the chromosomal anomaly

Hybrid gene

Description

5' BCL1 translocated on chromosome 14 near JH (junctions genes of IgH) and C in 3'; the breakpoint in BCL1 is in MTC (major translocation cluster), centromeric to the gene (in 5'), in 80% of cases, or dispersed in mTC1, 2, or 3 in 5' of the gene or in the 3' untranslated region of exon 5.

Fusion protein

Description

No fusion protein, but promoter exchange; the immunoglobulin gene enhancer stimulates the expression of BCL1.

Oncogenesis

Overexpression of BCL1 accelerates passage through the G1 phase.

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